**CASE REPORT**

**C2 Primary leiomyoma in an immunocompetent woman: A case report and review of literature**

Mohana Rao Patibandla, Madhukar T. Nayak, A. K. Purohit, Megha Uppin\(^1\), Sundaram Challa\(^1\), Gokul Chowdary Addagada\(^2\), Manisha Nukavarapu\(^2\)

Departments of Neurosurgery and \(^1\)Pathology, Nizam’s Institute of Medical Sciences, Hyderabad, Telangana, \(^2\)Guntur Medical College, Guntur, Andra Pradesh, India

**ABSTRACT**

Clinical case report and review of the literature. This is the first case of primary leiomyoma in an immunocompetent woman without previous history of uterine leiomyoma being reported in the literature to the best of our knowledge. Leiomyoma, a type of smooth muscle cell tumor, involving the vertebra is extremely rare. There were very few primary leiomyoma in patients with AIDS or in the immune-suppressed patients. This 48-year-old female came with H/o neck pain, weakness and bladder retention. On examination, tone increased in all four limbs, power on the right side of the limbs 4/5, power on the left upper limb 0/5, lower limb 3/5, left plantar was up going, decreased sensation over the left second cervical vertebra (C2) dermatome and all modalities decreased below C2. X-ray and magnetic resonance imaging (MRI) of the cervical spine showed kyphosis of the cervical spine with destruction of the C2 vertebral body along with pathological fracture. The patient underwent decompression of the C2 lesion through the C2 right pedicle with occipito-C1-C3 lateral mass screws fixation. Lesion anterior to the cord was reached by a transpedicular approach and decompression was performed. The lesion was pinkish grey, firm and moderately vascular and was destroying the C2 vertebral body. The patient improved symptomatically in power in the left upper limb and lower limb over the next 1 week duration from 0/5 to 4+/5. Histopathology revealed primary leiomyoma. The patient was evaluated with ultrasound abdomen and contrast tomogram of the chest, abdomen and pelvis to rule out other possible lesions in the lung, intestines and uterus. We suggest that leiomyoma should be included in the differential diagnosis of destructive lytic lesions involving the C2 vertebra. Histopathological examination with immunohistochemistry is necessary for the definitive diagnosis. Treatment of choice is surgery with complete removal.

**Key words:** Acquired immunodeficiency syndrome, benign metastasizing leiomyoma, immunocompetent, primary leiomyoma

**Introduction**

Primary smooth-muscle tumors of the central nervous system (CNS) are exceedingly rare. We report a primary extradural leiomyoma involving the second cervical vertebra (C2). There are many published reports of benign metastasizing leiomyoma (BML),\(^1\) but primary extradural leiomyoma is rarely reported.\(^2,3\) Primary leiomyoma of the bone is more common in the facial bones, and is less common in the skull bones and vertebra.\(^4\) Our report appears to be the first documentation of primary leiomyoma involving the C2 vertebra in an immunocompetent woman without an uterine leiomyoma. We also discuss the histogenesis and review the literature on smooth muscle tumors of the CNS.

**Case Report**

This 48-year-old female came with insidious-onset gradually progressive, continuous, dull aching type of pain on the right side of the neck radiating to the vertex region for 3 months. H/o weakness of all the four limbs along with tingling and numbness (Left > Right) were noted for 1 week, which...
was associated with bladder retention for 3 days. Motor examination showed increased tone in all four limbs. Power on the right upper limb was 4/5, power on the left upper limb was 0/5, lower limbs bilaterally was 3/5, left plantar was up going, sensory examination revealed decreased sensation over the left C2 dermatome and all modalities decreased below C2. X-ray of the cervical spine showed osteolytic destruction of the C2 body with subluxation of the C1 and C2 complex over C3 [Figure 1]. Magnetic resonance imaging (MRI) of the cervical spine showed kyphosis of the cervical spine with destruction of the C2 vertebral body along with pathological fracture. MRI also showed that the C2 vertebral body was showing T1 hypointense, T2 hyperintense lesions with posterior displacement of the severely compressed spinal cord [Figure 2]. Differential diagnosis of the expansive C2 vertebral body in immunocompetent and immunodeficiency patients is enumerated in Table 1. Preoperatively, our presumptive diagnosis was C2 vertebral body tuberculosis. The patient underwent decompression of the C2 lesion through the C2 right pedicle with occipito-C1-C3 lateral mass screws fixation (vertex

Figure 1: X-ray cervical spine showed osteolytic destruction of C2 body with subluxation of C1 and C2 complex over C3

Figure 2: Magnetic resonance imaging cervical spine showed kyphosis of the cervical spine with destruction of C2 vertebral body along with posterior displacement of the severely compressed spinal cord

Figure 3: X-ray showing occipito-C1-C3 lateral mass screws fixation with vertex Medtronic system
medtronic system) [Figure 3]. Lesion anterior to the cord was reached by a transpedicular approach and decompression was performed. The lesion was pinkish grey, firm and moderately vascular and was destroying the C2 vertebral body. Preoperative and intraoperative opinion was tuberculoma; therefore, safe maximal resection was performed instead of complete removal. The patient improved symptomatically in power in the left upper limb and lower limb over the next 1 week duration from 0/5 to 4+/5. Histopathological examination of the tumor tissue showed spindle cells arranged in whorls and fascicles with proliferation of smooth muscle cells surrounding the blood vessels. Spindle cells showed eosinophilic cytoplasm with elongated nuclei with blunt ends. There was moderate cellularity, minimal atypia, inconspicuous mitosis, and no evidence of necrosis; (c and d) Immunohistochemistry of the tumor cells stained positively for smooth muscle actin, desmin and vimentin and negative for epithelial membrane antigen, S-100, Cluster of differentiation 34, estrogen and progesterone receptors, confirming the diagnosis of the leiomyoma [Figure 4c and d] and excluding the other possible lesions like meningioma and
d

| Table 1: Differential diagnosis of the C2 vertebral body expansile lesion |
|-----------------------------|-----------------|-----------------|-----------------|
| Lesion                        | Age                     | Histology                                   | Imaging                                      |
| Hemangioma                   | any age; peak fourth decade | vascular spaces lined by endothelial cells | vertical parallel densities spotted appearance on CT high signal on T1W and T2W images; involvement of posterior elements |
| Langerhans cell histiocytosis | first, second decades | sheets of Langerhans cells, lymphocytes, and eosinophils | lytic lesion of the vertebral body leading to collapse |
| Aneurysmal bone cyst          | young patients upto 20 years vertebral body 40% | cystic spaces containing blood products | lytic expansile lesion with fluid-filled levels |
| Osteosarcoma                  | Fourth decade | Osteoid within sarcomatous tissue | Osteosclerotic and osteolytic areas with soft tissue component |
| Chondrosarcoma                | Fifth decade Predilection for vertebral body | Hyaline cartilage with increased cellularity within myxoid matrix | Bone destruction with characteristic puncate calcifications |
| Plasmocytoma                  | >40 years old | Sheets of plasma cells on a delicate reticular stroma | Radiolucent areas or reduction in bone density hypointense on T1W and hyperintense on T2W images |
| Giant cell tumor              | Third decade | Osteoclastic giant cells intermixed with spindle cells | Osteolytic geographic area with soft tissue component |
| Ewing's sarcoma               | Second to third decades | Sheets of small round blue cells | Lytic lesion, associated soft tissue mass |
| Chordoma                      | Middle-aged patients Exclusively affects vertebral body, most often sacrum, rarely mobile spine | Lobulated mass with mucinous containing cells | Destructive midline expansile lesion with associated soft tissue mass; extension into adjacent vertebra |
| Tuberculosis and vertebral Osteomyelitis | Younger age with or without Immunosuppression | Giant cells, lymphocytes, epithelioid cells | T1 Hypo, T2 hyper and enhances with contrast along with periepidural soft tissue |

CT – Computed tomography
Leiomyoma usually presents with myelopathy as in our case due to compression over the cord or radiculopathy due to compression of the nerve root. On X-ray or computerized tomogram (CT) imaging, leiomyoma of the vertebral body presents with osteolytic lesion with destruction of the vertebral body. On MRI, the tumor appears T1 isointense to hypointense and T2 hyperintense and enhances with contrast, as in our case. Presence of spindle cells with intense positivity for smooth muscle actin confirmed the diagnosis of leiomyoma.

The confusion in the diagnosis of the leiomyosarcoma and BML has been present for years; our tumor differs from leiomyosarcoma in view of absence of the mitoses, cellular atypia and necrosis. Therefore, it is suggested that the meticulous sampling of the pathology specimen, which indicates benign tendency, should not be easily diagnosed as leiomyosarcoma in spite of an aggressive course. The final diagnosis requires immunohistochemical staining and careful histological examination. The histological characteristics of this pathological entity differentiate it from other commonly occurring lesions like meningioma, hemangioma and metastasis. Thus, leiomyoma is one of the differential diagnoses for extradural lesions of the spine. The present neoplasm, which was diagnosed in C2 vertebra, had disclosed little histological signs of malignant character.

Treatment of leiomyoma in immunodeficiency states is same as in traditional primary leiomyomas. In case of the BML, they may be estrogen and progesterone receptor positive, and they have a tendency to recur after treatment. Therefore, medical treatment with leuteinizing hormone releasing hormone analogues and tamoxifen, or surgical treatment with a bilateral ophorectomy, may be useful. Shrinkage of the tumor after menopause and pregnancy, and even spontaneously, are reported with BML. These tumors are benign but have the potential to metastasize; therefore, complete removal of the tumor is needed.

**Conclusion**

Primary leiomyoma in the spine is very rare, and all previous reports are in immune-deficient patients. This is the first case report of the leiomyoma in the immunocompetent patient to the best of our knowledge. We recommend this diagnosis in differential diagnosis whenever an osteolytic spine lesion is noted. The treatment of choice is surgery with complete removal.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**